

1 1. A substantially pure mannin-binding lectin  
2 associated serine protease-2 (MASP-2) polypeptide.

1 2. The polypeptide of claim 1, said polypeptide  
2 being capable of associating with mannan-binding lectin  
3 (MBL).

1 3. The polypeptide of claim 1, said polypeptide  
2 being conjugated to a label or toxin.

1 4. A polypeptide containing the sequence  
2 identified as SEQ ID NO. 1.

1 5. A polypeptide according to claim 4 with a  
2 molecular mass of 20K.

1 6. A polypeptide with a molecular mass of 52K and  
2 containing the sequence identified as SEQ ID NO 1.

1 7. The polypeptide of claim 1, said polypeptide  
2 having serine protease activity.

1 8. A polypeptide of claim 1, said polypeptide being  
2 capable of MASP-2 activity in an *in vitro* assay for MBLectin  
3 complement pathway function.

1 9. A polypeptide according to claim 1, said  
2 polypeptide being capable of competitively inhibiting MASP-2  
3 serine protease activity.

1           10. A polypeptide according to claim 1 comprising a  
2 fragment of the polypeptide of SEQ ID NO:2, said polypeptide  
3 being a competitive inhibitor of complexing of MBL/MASP-2.

1            11. A polypeptide according to claim 5 or claim 6,  
2        said polypeptide having the amino acid sequence of SEQ ID  
3        NO:2.

1           12. A compound capable of competitively inhibiting  
2   serine protease activity of MASP-2 or a fragment thereof.

1           13. An isolated nucleic acid molecule of claim, the  
2 molecule comprising a nucleotide sequence encoding a  
3 polypeptide having sequence that is at least 85% identical  
4 to the sequence of SEQ ID NO. 1 or 2.

1           14. An isolated nucleic acid sequence encoding a  
2   mannan-binding lectin associated serine protease-2 (MASP-2)  
3   polypeptide according to claim 1.

1            15. A nucleic acid vector comprising the nucleic  
2    acid molecule of claim 14.

1           16. The nucleic acid vector of claim 15 wherein  
2   said vector is an expression vector.

1        17. The vector of claim 16, further comprising a  
2 regulatory element.

~~1 sub a 18. An antibody produced by administering an MASP-2~~  
~~2 polypeptide according to claim 1 to an antibody producing~~  
~~3 animal.~~

1 19. An antibody that selectively binds to MASP-2.

1 20. The antibody of claim 18 or claim 19, wherein  
2 said antibody is a monoclonal antibody.

1 21. The antibody of claim 18 or 19, said antibody  
2 being coupled to a compound comprising a detectable marker.

546 a<sup>2</sup>  
1 ~~22. A pharmaceutical composition comprising the~~  
2 ~~polypeptide of claim 1 or the antibody of claims 18 or 19.~~

1 ~~23. A method for detecting mannin-binding lectin~~  
2 ~~associated serine protease-2 (MASP-2), said method~~  
3 ~~comprising:~~  
4 ~~(a) obtaining a biological sample;~~  
5 ~~(b) contacting said biological sample with a MASP-2~~  
6 ~~polypeptide specific binding partner that specifically binds~~  
7 ~~MASP-2; and~~  
8 ~~(c) detecting said complexes, if any, as an~~  
9 ~~indication of the presence of mannin-binding lectin~~  
10 ~~associated serine protease-2 in said sample.~~

1 ~~24. A method according to claim 23, in which the~~  
2 ~~specific binding partner is an antibody.~~

1 ~~25. A method for detecting MASP-2, said method~~  
2 ~~comprising an assay for MASP-2 complement fixing activity.~~

1 ~~26. The methods of claims 23 or 24 for quantitative~~  
2 ~~assay of MASP-2 or MASP-2 activity in biological samples.~~

1 27. A method for detecting MASP-2 nucleic acid  
2 expression, comprising detecting RNA having a sequence  
3 encoding a MASP-2 polypeptide by mixing the sample with a  
4 nucleic acid probe that specifically hybridizes under  
5 stringent conditions to the nucleic acid of claim 13 or 14.

1 28. A method for treating patients deficient in  
2 MASP-2 by administering to the patient the peptide of claim  
3 1.

1 29. A method for treating patients deficient in  
2 MASP-2 by administering to the patient nucleic acid  
3 according to claim 13 or 14.

1 30. A method for inhibiting the activity of MASP-2  
2 by administering to the subject a compound that inhibits  
3 expression or activity of MASP-2.

1 31. The method of claim 27 in which the compound is  
2 a MASP-2 anti-sense nucleic acid sequence.

1 32. The method of claim 30 comprising administering  
2 a compound that inhibits complexing of MBL and MASP-2.

1 33. An assay for polymorphisms in the nucleic acid  
2 sequence encoding MASP-2.

1 34. A method of detecting the presence of MASP-2-  
2 encoding nucleic acid in a sample, comprising mixing the  
3 sample with at least one nucleic acid probe capable of  
4 forming a complex with MASP-2-encoding nucleic acid under  
5 stringent conditions, and determining whether the probe is  
6 bound to sample nucleic acid.

1 35. A nucleic acid probe capable of forming a  
2 complex with MASP-2-encoding nucleic acid under stringent  
3 conditions.

1 36. An assay for polymorphisms in the polypeptide  
2 sequence comprising MASP-2 or its precursor.

1 37. A method for diagnosing a disorder associated  
2 with aberrant expression of MASP-2, comprising obtaining a  
3 biological sample from a patient and measuring MASP-2  
4 expression in said biological sample, wherein increased or  
5 decreased MASP-2 expression in said biological sample  
6 compared to a control indicates that said patient suffers  
7 from a disorder associated with aberrant expression of MASP-  
8 2.

1 38. A method for diagnosing a disorder associated  
2 with aberrant activity of MASP-2, comprising obtaining a  
3 biological sample from a patient and measuring MASP-2  
4 activity in said biological sample, wherein increased or  
5 decreased MASP-2 activity in said biological sample compared  
6 to a control indicates that said patient suffers from a  
7 disorder associated with aberrant activity of MASP-2.

1           39. A method of assaying for activity MBL-complexed  
2   MASP, the method comprising  
3           providing a sample to be assayed and substantially  
4   reducing any artifact resulting from activation of the  
5   classical complement fixing pathway by conducting the assay  
6   in the presence of an ionic strength high enough to  
7   effectively reduce activation of the classical complement  
8   fixing pathway but not so high as to substantially interfere  
9   with activity of MBL-complexed MASP.

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